IN THE CLAIMS:

This listing of claims replaces all prior versions and listings of claims in the application:

Listing of Claims

1-25. (Canceled)

26. (Previously presented) A transgenic *D. melanogaster* comprising

a transgene containing a plurality of CAG's and at least one CAA sequence encoding a polyglutamine repeat sequence operably linked to a constitutive, regulatable, or tissue specific expression control element, wherein the tissue specific expression control element is selected from the group consisting of Appl, rhodopsin 1 promoter, and GLASS transcription factor element, and wherein the transgene produces polyglutamine toxicity in the transgenic *D. melanogaster*.

27-28 (Canceled)

29. (Previously Presented) The *D. melanogaster* of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 1:1 and 2:1.

30. (Previously Presented) The *D. melanogaster* of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 2:1 and 5:1.

31. (Previously Presented) The *D. melanogaster* of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 5:1 and 10:1.

32. (Previously Presented) The *D. melanogaster* of claim 26, wherein the number of CAG's to CAA's is in ratio of between about 10:1 and 50:1.

33. (Canceled)

34. (Previously Presented) The *D. melanogaster* of claim 26, wherein the tissue specific expression control element confers neural, retinal, muscle or mesoderm cell expression.

35-36. (Canceled)

37. (Previously Presented) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about 50 and 100 amino acids in length.

38. (Previously Presented) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about 100 and 200 amino acids in length.

39. (Previously Presented) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence is between about 50 and 200 amino acids in length.

40. (Previously Presented) The *D. melanogaster* of claim 26, wherein the polyglutamine sequence further comprises a tag.

41. (Canceled)

42. (Currently Amended) The *D. melanogaster* of claim 26, wherein the Drosophila further comprises a marker sequence inserted into its genomic DNA, wherein the marker is located adjacent to a gene selected from the group consisting of a HDJ1 gene, a TPR2 gene, and a MLF gene ~~a gene or inserted into a gene whose expression or activity increases or decreases polyglutamine toxicity~~ in the animal, and wherein the marker sequence comprises an inducible upstream activating sequence, a minimal promoter sequence and 5′ and 3′ P transposon elements containing terminal inverted repeats.

43. (Canceled)

44. (Currently Amended) The *D. melanogaster* of claim 42 ~~43~~, wherein the gene is HDJ1.

45. (Currently Amended) The *D. melanogaster* of claim 42 ~~43~~, wherein the gene is TPR2.

46. (Currently Amended) The *D. melanogaster* of claim 42 ~~43~~, wherein the marker sequence is near an MLF gene.

47. (Withdrawn) A method for identifying a compound that modulates polyglutamine toxicity in an animal comprising:

(a) contacting the animal of claim 41 with a test compound; and

(b) determining whether the test compound increases or decreases polyglutamine toxicity in the animal, where increased or decreased polyglutamine toxicity identifies the test compound as a compound that modulates polyglutamine toxicity.

48. (Withdrawn) The method of claim 47, wherein the compound is present in the animal's food or drink.

49. (Withdrawn) The method of claim 47, wherein the compound is administered to a tissue or organ of the animal.

50. (Previously Presented) A method of producing a transgenic *D. melanogaster* characterized by suppressed polyglutamine toxicity comprising:

(a) transforming a *D. melanogaster* embryo or fertilized egg with a transgene comprising a plurality of CAA and CAG sequences encoding a polyglutamine sequence operably linked to a constitutive, regulatable, or tissue specific expression control element, wherein the tissue specific expression control element is selected from the group consisting of Appl, rhodopsin 1 promoter, and GLASS transcription factor element and

(b) selecting a *D. melanogaster* that exhibits polyglutamine toxicity.

51. (Withdrawn) An isolated polynucleotide sequence having about 65% or more identity to a Drosophila TPR2 (dTPR2) sequence set forth as SEQ. ID NO:2 and which encodes a polypeptide that decreases polyglutamine toxicity, with the proviso that the

sequence is distinct from the EST sequences set forth in Figure 11.

52. (Withdrawn) The polynucleotide sequence of claim 51, wherein the sequence encodes a subsequence of TPR2 that decreases polyglutamine toxicity.

53. (Withdrawn) The polynucleotide sequence of claim 51 operatively linked to an expression control element.

54. (Withdrawn) An isolated polynucleotide sequence that hybridizes under stringent conditions to a Drosophila TPR2 (dTPR2) sequence set forth as SEQ. ID NO:2, with the proviso that the sequence is distinct from the EST sequences set forth in Figure 11.

55. (Withdrawn) The polynucleotide sequence of claim 54, wherein the sequence comprises a polynucleotide having 20 or more contiguous nucleotides.

56. (Withdrawn) The polynucleotide sequence of claim 54, wherein the sequence comprises a polynucleotide having 50 or more contiguous nucleotides.

57. (Withdrawn) An isolated polynucleotide sequence having about 65% or more identity to a Drosophila MLF (dMLF) sequence set forth as SEQ. ID NO:4 and which encodes a polypeptide that decreases polyglutamine toxicity, with the proviso that the sequence is distinct from the EST sequences set forth in Figure 12.

58. (Withdrawn) The polynucleotide sequence of claim 57, wherein the sequence encodes a subsequence of MLF that decreases polyglutamine toxicity.

59. (Withdrawn) The polynucleotide sequence of claim 57 operatively linked to an expression control element.

60. (Withdrawn) An isolated polynucleotide sequence that hybridizes under stringent conditions to a Drosophila MLF (dMLF) sequence set forth as SEQ. ID NO:4, with the proviso that the sequence is distinct from the EST sequences set forth in Figure 12.

61. (Withdrawn) The polynucleotide sequence of claim 60, wherein the sequence comprises a polynucleotide having 20 or more contiguous nucleotides.

62. (Withdrawn) The polynucleotide sequence of claim 60, wherein the sequence comprises a polynucleotide having 50 or more contiguous nucleotides.

63. (Withdrawn) A composition comprising a polynucleotide sequence encoding a human MLF polypeptide operatively linked to an expression control element in a pharmaceutically acceptable carrier.

64. (Withdrawn) A composition comprising a polynucleotide sequence encoding a human TPR2 polypeptide operatively linked to

an expression control element in a pharmaceutically acceptable carrier.

65. (Withdrawn) A method of increasing survival of a cell having polyglutamine toxicity, comprising contacting the cell with an amount of TPR2 or MLF polypeptide sequence or a polynucleotide sequence TPR2 or MLF polypeptide to increase survival of the cell.

66. (Withdrawn) A method of decreasing apoptosis of a cell, comprising contacting the cell with an amount of TPR2 or MLF polypeptide sequence or a polynucleotide sequence TPR2 or MLF polypeptide to decrease apoptosis of the cell.

67. (Withdrawn) A method of decreasing polyglutamine toxicity in a cell having or susceptible to polyglutamine toxicity, comprising contacting the cell with an amount of J domain containing polypeptide, TPR2 or MLF polypeptide sequence, or a polynucleotide sequence encoding the J domain containing polypeptide, TPR2 or MLF polypeptide sequence to decrease polyglutamine toxicity in the cell.

68. (Withdrawn) The method of claim 67, wherein the cell is a neural, retinal, muscle or mesoderm cell.

69. (Withdrawn) The method of claim 67, wherein the toxicity is decreased by decreasing cell death or increasing cell survival.

70. (Withdrawn) A method of decreasing polyglutamine toxicity in a tissue or organ of a subject having or at risk polyglutamine toxicity, comprising contacting the tissue or organ with an amount of a J domain containing polypeptide, a TPR2 or MLF polypeptide sequence, or a polynucleotide sequence encoding the J domain containing polypeptide, TPR2 or MLF polypeptide, to decrease polyglutamine toxicity in the tissue or organ of the subject.

71. (Withdrawn) The method of claim 70, wherein the tissue is brain, eye, muscle or mesoderm.

72. (Withdrawn) A method of decreasing the severity of a frontotemporal dementia, prion disease, polyglutamine disorder or protein aggregation disorder in a subject having or at risk of a frontotemporal dementia, prion disease, polyglutamine disorder or protein aggregation disorder, comprising administering to the subject an amount of J domain containing polypeptide, a TPR2 or MLF polypeptide sequence, or a polynucleotide sequence encoding the J domain containing polypeptide, TPR2 or MLF polypeptide, to decease the severity of the frontotemporal dementia, prion disease, polyglutamine disorder or protein aggregation disorder in the subject.

73. (Withdrawn) The method of claim 72, wherein the method comprises prophylactic administration.

74. (Withdrawn) The method of claim 72, wherein the disorder is a neurological or muscle disorder.

75. (Withdrawn) The method of claim 72, wherein the disorder impairs long term or short term memory or coordination of the subject.

76. (Withdrawn) The method of claim 72, wherein the disorder is characterized by the presence of protein aggregates, amyloid plaques, degeneration or atrophy in an affected tissue or organ.

77. (Withdrawn) The method of claim 72, wherein the disorder is selected from the group consisting of Alzheimer's disease, Parkinson's disease, Creutzfeldt-Jacob's disease (CJD), bovine spongiform encephalopathy, Huntington's disease (HD), Machado-Joseph disease (MJD), Spinocerebellar ataxias (SCA), dentatorubropallidoluysian atophy (DRPLA), Kennedy's disease, stroke and head trauma.

78. (Withdrawn) The method of claim 72, wherein the severity is decreased by decreasing cell death or increasing cell survival.

79. (Withdrawn) The method of claim 72, wherein the severity is decreased by decreasing protein aggregation.